

# Genital Surgery for Disorders of Sex Development: Implementing a Shared Decision-Making Approach

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## ABSTRACT

Ongoing controversy surrounds early genital surgery for children with disorders of sex development, making decisions about these procedures extraordinarily complex. Professional organizations have encouraged healthcare providers to adopt shared decision-making due to its broad potential to improve the decision-making process, perhaps most so when data are lacking, when there is no clear “best-choice” treatment, when decisions involve more than one choice, where each choice has both advantages and disadvantages, and where the ranking of options depends heavily on the decision-maker’s values. We present a 6-step model for shared decision-making in decisions about genital surgery for disorders of sex development: 1) Set the stage and develop an appropriate team; 2) Establish preferences for information and roles in decision-making; 3) Perceive and address emotions; 4) Define concerns and values; 5) Identify options and present evidence; and 6) Share responsibility for making a decision. As long as controversy persists regarding surgery for DSD, an SDM process can facilitate the increased sharing of relevant information essential for making important health care decisions.

## KEY WORDS

(MeSH): decision-making; sex differentiation disorders; Hermaphroditism; urogenital surgical procedures

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## ABBREVIATIONS

CAH—Congenital adrenal hyperplasia  
DSD—Disorders of sex development  
RCT—Randomized controlled trial  
SDM—Shared decision-making

## INTRODUCTION

Disorders of sex development (DSD) is an umbrella term adopted to refer to the host of “congenital conditions in which development of chromosomal, gonadal, or anatomical sex is atypical”<sup>1</sup>. For those DSD characterized by atypical genital appearance, standard practice since the 1950s has been to perform surgery intended to modify the infant’s genitals to make them more typical for the sex of rearing to facilitate gender identity development<sup>2-5</sup>. For more than a decade, however, there has been intense debate both within and outside medicine regarding the timing, necessity, benefits, and risks of elective genital surgery for children born with DSD<sup>6</sup>.

(There is no dispute about the necessity of operations to address imminent threats to the infant’s health, such as the creation of a urinary opening.)

Decisions about genitoplasty are made challenging by the dearth of clinical evidence regarding long-term psychological and physical outcomes<sup>1,6</sup>. Because evidence is lacking (and existing evidence is contested)<sup>1,7</sup>, providers make treatment recommendations based in large part on their personal values, anecdotal evidence, and “gut feeling”<sup>13-16</sup>. Some providers further report feeling unsure about the “right” decision in such cases<sup>8</sup>. Meanwhile, some parents express dissatisfaction with the information they receive

about surgical options, report feeling rushed into a decision, and express decisional regret<sup>8</sup>. Taken together, these circumstances make decisions about genital surgery extraordinarily complex.

In recent years, organizations such as the Institute of Medicine, the Agency for Healthcare Research and Quality, the American College of Physicians, the American Academy of Family Physicians, the Society of Critical Care Medicine, and the American Urological Association have encouraged doctors to adopt a shared decision-making (SDM) approach when complex or difficult decisions must be made regarding interventions or appropriate goals of care<sup>17-23</sup>. SDM is an important tool for making health care decisions and this is perhaps most so when data are lacking, when there is no clear “best-choice” treatment, when decisions involve more than one choice, where each choice has both advantages and disadvantages, and where the ranking of options depends heavily on the decision-maker’s values<sup>9,10</sup>. Despite SDM’s promise, the lack of specific models outlining how to implement SDM in particular contexts has meant its adoption has been uneven<sup>10</sup>.

In this paper, we present a 6-step model for the application of SDM in decisions about genital surgery for children born with DSD. Although SDM cannot provide a “right” answer in such difficult situations, an SDM process can facilitate the increased sharing of relevant information essential for making important health care decisions. SDM gives providers the opportunity to clarify the basis for their recommendations, identify and fill gaps in parents’ understanding about the diagnosis and treatment options, and explore the values underlying both parents’ and clinicians’ concerns. The process of thoroughly examining alternatives and the encouragement of transparency and questioning involved in SDM can help to ensure that the best interests of the child and family are served, patient care and the doctor-patient relationship are improved, satisfaction with the decision-making process is increased for both physician and parents, decisional conflict and regret are minimized, and ethical and legal requirements for informed permission (and assent when appropriate) are met.

## WHAT IS SHARED DECISION-MAKING?

SDM is an evolving concept, but generally centers on the collaboration of patients and caregivers in making difficult healthcare choices allowing them jointly to bear the burden and responsibility of decision-making<sup>9,11-14</sup>. SDM differs in several key ways from other widely cited approaches to medical decision-making such as the paternalistic model and the autonomy-driven (or informed) model. In the paternalistic model, the physician is understood as best suited to determine the patient’s best interest and hence the patient is given limited information and a passive role in the treatment decision-making process largely limited to providing consent for the treatment the physician recommends.<sup>14</sup> This model was dominant in the United States through the 1960s and remains the standard in many parts of the world.<sup>15</sup> The autonomy-driven (or informed) decision-making model places the authority to decide squarely on the patient herself, and under such a model the physician primarily provides the patient with information about her medical condition and treatment options. In the autonomy-driven model, which has become the standard in the United States<sup>15</sup>, the physician’s role is largely one of information sharing<sup>11</sup>. This model, however, ignores data showing that most patients prefer a shared decision-making process, and thus autonomy-driven decision-making often leaves patients (and presumably parents) feeling abandoned by the physician<sup>16-20</sup>. Data also suggest that surgeons may be more directive than medical physicians<sup>21</sup>; therefore, without an SDM model, surgeons may have more influence on parental decisions than non-surgeons, leading to higher rates of surgery<sup>22</sup>.

By contrast, SDM is a *process*; it necessarily requires clinical caregivers to reveal their reasoning, values, and biases, and to similarly explore the understandings, values, and reasoning of patients or their surrogates<sup>9,10,23,24</sup>.

SDM can also help caregivers meet legal and ethical standards for informed consent (or informed permission—with the assent of the child as appropriate—in the case of pediatric patients), which generally require physicians to provide all

information a reasonable parent (and child when appropriate) would find material in making an informed decision, and to fully inform parents of all options available regardless of the provider's biases or preferences<sup>25-27</sup>. SDM allows caregivers and parents to engage in a thoughtful discussion of the pros and cons of treatment options by helping to uncover the values and beliefs framing the caregiver's recommendations<sup>28</sup> and the parents' decisions. SDM is not only likely to create more realistic expectations and reduce the decisional conflict associated with feeling uninformed, but it can help to elicit the "best option" for decisions involving close tradeoffs that can only be made by incorporating personal preferences<sup>9,10</sup>.

### A MODEL FOR SHARED DECISION-MAKING

In what follows we outline a process for SDM in the context of decisions about elective genitoplasty for infants and young children with DSD. Given the challenges of making decisions about genitoplasty, a structured process can be useful to ensure that parents truly share in decision-making<sup>29</sup>. The steps outlined below seek to address parents' emotional and informational needs, and allow for reflection and transparency during decision-making. In practice, discussions may not always fall neatly into the six sequential steps. It is important to note that while parents and clinicians may wish to attend to genital atypicality as quickly as possible once the child has been assigned a gender (and surgical possibilities may inform gender assignment decisions), gender assignment is a social and legal process that may proceed irrespective of medical or surgical intervention<sup>1</sup>.

#### 1. Set the stage and develop an appropriate team

For a complex decision like elective genitoplasty in cases of DSD, preparation for the SDM process begins well before the clinical consultation. It is important to consider the range of expertise needed, how to frame the decisions to parents, and how to enhance parents' understanding of the decisions they face. A balanced presentation of information may require the

provider to consider in advance how to discuss matters that might have gone unaddressed in the past, or how to examine unspoken assumptions. Examples below will highlight some of these areas for extra consideration. The *Consensus Statement on Management of Intersex Disorders* specifies that "optimal care for children with DSD requires an experienced multidisciplinary team"<sup>1, e490</sup> comprised of a range of pediatric subspecialists (e.g., endocrinology, urology/surgery, psychology/psychiatry, gynecology, social work, nursing, bioethics, and child life). The team should also include the child's pediatrician (or other primary medical provider) who can bring a whole-child/whole-family perspective and who may help to facilitate collaboration, especially if disagreements arise over decision-making and patient "ownership"<sup>1,30</sup>. Moreover, because pediatric specialists do not generally provide care to patients with DSD beyond adolescence, input from specialists such as reproductive endocrinologists and urologists who provide care to adults with DSD may be critical to understanding long-term sequelae of pediatric treatments.

Although a team concept has been advanced as an ideal to transform DSD care, it may be perceived as a threat to existing hierarchies, and thus could create conflict and discomfort for caregivers who are not empowered in the current order. Education may be an important part of the transition to team care and enlightened leadership on the part of those in power will help to increase the feasibility of a team model<sup>31</sup>.

Communication is a central piece of SDM and a provider with appropriate communication skills will need to be identified as the family's primary liaison, ensuring that parents are involved in team discussions and connected to resources and support. Advances in fields such as oncology may prove useful for improving the communication skills of practitioners in the field of DSD. The National Cancer Institute, for example, has identified six core functions of cancer communication that parallel those we are suggesting<sup>32</sup> and a number of innovative programs and methods (e.g., trigger videos, patient and provider stories, self-reflection, role-play, and supervised practice with standardized patients) have been developed to improve caregivers' patient-

centered communication skills<sup>33-36</sup>.

Once a team has been identified, it will need to reach a unified understanding of the goals of treatment and the reasonable alternatives. Furthermore, it should ascertain the family's full range of needs and how best to meet those needs and to integrate care. As much as possible, the team should include parents, and the child when appropriate, as partners in the discussions regarding the child's care. Involvement of parents early in the process (in some cases before recommendations have been developed) will help parents understand the full range of perspectives, will help providers understand the values and concerns of the family, and will create transparency. Mutual respect, open discussion, and the valuing of multiple perspectives will support the broad participation of the parents and other team members in discussions and problem-solving. Team members will need to work to ensure parents' views and needs are made explicit and that caregivers are responsive to those needs. Families should further understand that they may have options regarding who is on the care team, and that they are empowered to discharge providers or to seek care from other providers if they wish.

## **2. Establish preferences for information and roles in decision-making**

Early in discussions, it is important to ask parents about their preferences for information and to discuss the role of all parties in making a decision. It is important to address these matters explicitly to minimize the risk of physicians making unilateral decisions and incorrect assumptions about patients' information needs and preferences<sup>37-40</sup> or incorrectly assessing their own information-giving behavior<sup>28</sup>. Some parents will want the caregivers to take the role of "unbiased educators"<sup>41</sup>, whereas others will prefer caregivers be active participants in making a decision<sup>42</sup>. Parents may have different preferences about the depth of coverage for some kinds of information; however there is consistent evidence that patients often want a greater degree of information than they receive, even when they want their physician to make important healthcare decisions<sup>43-45</sup>. In the case of DSD, physicians may

be tempted to give parents incomplete or inaccurate information about sensitive topics like sexual development or future sexual function in the belief that parents "can't handle such information."<sup>8</sup> Such practices violate accepted norms of informed permission,<sup>46, 47</sup> and may overstep the training of healthcare providers who may not be adequately equipped to judge what parents can or cannot handle.<sup>48</sup>

## **3. Perceiving and addressing emotions**

It is difficult to make rational decisions when parents' strong emotional responses remain unacknowledged. Research on parents' experiences at the birth of their child with a DSD—when surgical decisions are first broached—shows they typically experience shock, confusion, fear, guilt, anger, sadness, anxiety, shame and alienation, and may also grieve the loss of their anticipated "perfect child"<sup>8,49,50</sup>. These findings are consistent with findings from parents of children with other congenital anomalies<sup>51,52</sup>. It is thus important for team members to take great care in relaying information to the parents so as to "get it right from the start"<sup>53</sup>. This includes reassuring parents through words and attitude that these feelings are normal and acceptable, to emphasize that the child's condition is only one aspect of the child, to stress the positive aspects of their newborn child, and to ensure that decisions are not made before these emotional responses have been explored<sup>53-55</sup>. This will require providers not only to identify parents' emotions, but also to recognize their own emotions and reactions including how their words and behavior may affect parents<sup>56</sup>. "For example, clinicians may feel embarrassment at not having all the answers, fear that the parents may make what the practitioner believes is the 'wrong' decision, or a desire to 'save' the parents from their discomfort or the child from a difficult future."

Clinicians sometimes recommend genitoplasty to assuage parental anxiety, but this approach thwarts shared decision-making. Parents who are overly anxious about the child's atypical genitals may not be prepared to effectively participate in the decision-making process and may be unable to offer truly *informed* permission<sup>46</sup>. To the extent

possible, parents who are experiencing strong emotional responses need help addressing these feelings before authorizing elective surgery. This is just one example where the presence of a behavioral health expert on the team can be helpful.

Traditionally, treatment for infants with DSD has centered on interventions aimed at eliminating atypical sex characteristics based on the belief that doing so will ease the child's adjustment and relieve parental distress<sup>6</sup>. However, the Consensus Statement on Management of Intersex Disorders notes that evidence to support this belief is lacking<sup>1</sup>. Data indicate that parents' concerns about their child's gender-atypical biology (and even genitals) persist long after genital surgery and evolve with new developmental stages<sup>8</sup>. Exclusive focus on immediate surgical intervention will thus fail to address the broad range of issues a family will face, even if surgeons or the family deem surgery successful<sup>8</sup>. Similarly, parents who decide to forego surgery may experience new or heightened concerns as the child develops, which will need to be addressed.

For these reasons, the team will need to take a broad and subjective view of the complex and evolving needs of the family and patient focusing on both the child's short- and long-term needs and well-being<sup>1</sup>. A whole-child/whole-family perspective such as this would start with the questions: "What is the full range of issues the child's condition raises for the family and how can we address those?" "What are the child's most important needs, now and in the future?" "What are the family's most important needs, now and in the future?" Addressing these questions is likely to lead to a range of interventions, including education for the family, provision of psychological and support services, and connection with peer support groups.

#### 4. Defining concerns and values

Before discussing surgical options, it is useful to describe the diagnosis, when and if it is known, to parents and then to explore their related concerns. How physicians frame these discussions can have a significant impact on how the

family perceives the situation and their child. In the case of a DSD, if the problem is defined as "abnormal genitals," surgery may be the only obvious alternative to non-treatment. However, if the problem is defined as "potential for teasing" or "challenges of growing up with an atypical body," a range of other options becomes apparent such as psychosocial and peer support. To locate core concerns, it may be helpful to ask, "What problem is this causing?" which will shift the focus away from a choice of "surgery or no surgery" to "How do we best address the concerns we've identified?"

Once concerns have been identified (and recognizing concerns will change and evolve), it is important to explore the parents' values, which will help in weighing the pros and cons of different treatment options. What do *these* parents want for *this* child's life? What do they hope to get from treatment? Often, when there is no easy decision, it is because competing values are in play. For example, when parents are considering genital surgery for a child with a DSD, their values may include protecting the child from stigma, ensuring a "normal" childhood, avoiding unnecessary interventions, keeping surgical and gender options open for the future, ensuring the best physical outcome, ensuring the best psychological outcome, supporting the child's healthy sexual and gender identity development, enabling the child to have a healthy sex life in adulthood, and preserving fertility. If the team understands how the parents weigh these different values, it will be easier to outline treatment options in a way that addresses parents' concerns. Simply because parents do not raise an issue does not mean it is not a concern. For example, it may be difficult for some parents and even clinicians to discuss the future sexual pleasure of a newborn. However, it is critical that the issue is discussed to determine how parents value this, and to meet requirements for informed permission<sup>46</sup>. Table 1 presents an extensive list of issues to be considered in such discussions for infants with DSD.

Healthcare providers must explicitly articulate their thoughts about treatment options without being so forceful that their opinions overshadow those of the parents. Because physicians are in

positions of power, even mentioning their own judgment can become coercive if not done with great tact<sup>66,57,58</sup>. Further, while physicians have expert medical knowledge, this does not translate into expert knowledge of values or personal preferences<sup>57,59</sup>, therefore the provider must acknowledge to the parents that his or her values are not more “right” than theirs. Some practitioners, for example, may find it irresponsible or harmful to allow a child to grow up with genitals that are atypical for her gender assignment. Others may view atypical genitals as a manageable challenge, but may be concerned about making irreversible decisions early in the child’s life. Such value judgments are likely to weigh in a provider’s treatment recommendations. Because there are so few data on the long-term consequences of surgery on children and families, as well as for foregoing surgery, opinions are likely to be based on beliefs rather than on “facts”<sup>1,8</sup>. As such, clinicians must recognize the limitations of their own beliefs, and should help parents consider their own assumptions and biases. Doing so will enable parents to assess the extent to which their own values overlap with those of the provider and allow them to judge how to weigh the provider’s recommendations. A well-functioning multidisciplinary team can also mitigate any undue influence of an individual provider’s perspective<sup>30</sup>.

### 5. Identify options and present evidence

In an ideal SDM scenario, the primary liaison should identify and present all options to the parents (including the option of no surgery), the possible consequences of each option, and how likely the consequences are. The liaison should also reveal whether any proposed interventions represent the standard of care, and whether different practitioners would recommend other approaches and why<sup>28,60-63</sup>. This level of detail assumes physicians have precise information about the risks and benefits of each option. With DSD, however, much evidence is lacking about the extent to which surgery may improve quality of life, retain sexual sensation, or provide the psychosocial benefits for which parents and clinicians hope<sup>1</sup>. Similarly, we have scant

contemporary evidence about how a child and family may fare having foregone surgery. Where evidence is available, risks and benefits should be presented and evaluated in relation to the individual child and the parents’ concerns and values. Providers have a corresponding obligation to inform parents about the type and quality of evidence including gaps in, and limitations of, evidence<sup>47</sup>. Judgments about evidence are complex and require that providers assess the quality of that evidence. Assessments may include implicit judgments regarding which outcomes are most important, the quality of evidence for each important outcome, the overall quality of evidence, balance between benefits and harms, and value of incremental benefits. In presenting evidence, providers should take into account the hierarchy of studies for obtaining evidence: systematic reviews of randomized controlled trials (RCTs); evidence from at least one RCT; controlled observational studies such as cohort and case control studies; uncontrolled observational studies such as case reports; expert committee reports, expert opinion, or clinical experience. Expert opinion should not be confused with personal experience. In the absence of other forms of evidence, expert opinion may be the best guide available.

In presenting information, providers should strive for objectivity. The process of identifying options and presenting evidence will necessarily involve back-and-forth as parents may redefine values in the face of concrete options. The primary liaison must explain not only what the team believes is the best course of action, but whether this recommendation is supported by evidence and if not, on what basis this recommendation is being made<sup>47, 64</sup>.

### Be realistic

It is important that the liaison is realistic about the known risks and sequelae of surgery, such as pain, risk of complications, infection and even death as well as those uncertainties associated with foregoing surgery. Particularly for elective surgery with uncertain outcomes, these known hazards are relevant and need to be weighed<sup>46</sup>.

Parents often have unrealistic expectations

that surgery will “fix” their child, and this misunderstanding may be exacerbated by clinicians’ words. The surgeon Jeffrey Marsh has noted that surgeons tend to say they will “correct” a certain feature when “it is rarely the case that the operation is so perfect that there is no residue of the original difference”<sup>41</sup>. Clinicians should be cognizant that parents tend to over-inscribe the extent to which surgery may resolve their concerns.

Sometimes, a realistic perspective can best be obtained from someone with first-hand experience of a decision and its consequences. Existing models of SDM emphasize the importance of providing opportunities for parents to talk with others who have lived through positive and negative outcomes of various treatment options. Because parents of children with DSD frequently have no personal experience of their child’s condition, and because they may have to make initial decisions without the child’s input, it is especially important for them to have the opportunity to talk with people who have first-hand experience of living with DSD. Several national support groups exist which can facilitate such interaction<sup>1</sup>.

### **Explore parents’ ideas and assumptions, correct misperceptions**

Even after careful explanation, parents may make inaccurate assumptions about the need for or consequences of treatment. For example, parents may assume that genitoplasty will reduce the chances the child will grow up to be homosexual.<sup>49</sup> If unrealistic expectations are a basis for decision-making, parents need to know of the absence of evidence supporting their assumption. To identify these unfounded beliefs, the healthcare team may need to probe parents for the reasoning that underlies their decisions. Additionally, it is important to check whether parents understood the information presented to them. This may mean asking them to articulate their understanding of the child’s condition and the treatment options, and asking their reasons for leaning towards one or another alternative.

### **Ensure understanding**

Physicians should ensure that the information given to the parents is reasonable for them and that the parents understand the full range of options available as well as the known (and unknown) ramifications of those options<sup>47,60,65</sup>. Throughout the SDM process there should be ample opportunity for parents and clinicians alike to absorb information, ask questions, and explore and re-clarify needs, concerns, and values. It is not enough to assume that parents will ask questions if they don’t understand information, that they will speak up when they disagree with a physician’s reasoning or values, or that what parents hear is what the physician means to convey and vice versa. What is communicated and what is heard will be shaped by the clinicians’ and parents’ understanding, perceptions, and valuations of the child’s condition and the challenges they believe it to pose as well as the benefits and harms posed by the various treatment options. DSD pose particular challenges to parental understanding, since the subject is scientifically complex and unfamiliar to most people. Adding to this complexity, the concept of sex and gender difference is fraught with stereotypes, value judgments and emotional triggers<sup>29,46</sup>. It may be helpful to involve a behavioral health team member who has experience navigating emotionally fraught issues, ideally but not necessarily someone with specific knowledge of DSD.

### **6. Sharing responsibility for making a decision**

At the end of this process, a difficult decision must ultimately be made. But the *process* of SDM—going through the steps outlined above—helps to ensure that responsibility for that decision is shared in a meaningful way. The parents will have gained technical information and emotional support, which will help them to make sound choices. The team will have gained insight into the parents’ priorities and the family’s situation, which can inform recommendations. Often, an SDM process will result in a consensus built on trust and understanding.

Even with a good process, however, sometimes parents and providers will disagree on the best course of action. In such a case, it becomes clear that SDM means a shared *process*, not necessarily a shared *decision*. Parents and providers have shared responsibility, but different kinds of authority<sup>13,24</sup>. A physician is not required to provide treatment that is contra-indicated, no matter how strongly parents request it. Further, a physician may decline to participate in interventions when he believes participation would violate his or her professional or personal integrity. Parents need not authorize elective treatment they feel is not in the child's best interest, and they have the right to change providers.

Ultimately, providers must recognize that it is the child, and then the parents, who are most affected by the decision, and temper their input accordingly<sup>24</sup>. Because physicians and other care providers often have strong attachments to their patients and are responsible for the care and safety of the child, they are "stake-holders" in the decision-making process. However, because the providers do not share equally the burdens of the decisions made (e.g., they will not be the ones caring for the child on a day-to-day basis), it is the values of the parents (and the child when appropriate) that guide the decision-making process. Indeed, "without direct evidence that any one treatment is superior to another, parent and patients preferences regarding treatment options should generally be honored"<sup>64</sup>.

In some cases, parents and providers will decide to take no immediate action and continue the decision-making process over time. In order for the decision to truly be shared, parents need time to absorb information and explore their responses. Parents may benefit from a second opinion or from talking with other parents who have faced similar choices including those who chose surgery, those who chose a non-surgical option, those who are content with the choices they made, and those who regret their decisions. Furthermore, it may be necessary to revisit some of the steps repeatedly as parents process information and emotional responses, and gain experience living with their child's DSD.

## CONCLUSION

SDM as a tool has broad potential to improve the decision-making process. While our discussion has focused on infants with DSD, this model can be adapted to many other situations in pediatrics. By systematically sharing information, exploring concerns, and exposing the values that underlie parents' and providers' perspectives, SDM enables both parties to take active roles in the decision. This will be especially critical in DSD where much still remains contested and unknown regarding genital surgery.

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## DISCLOSURE

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## REFERENCES

1. Lee PA, Houk CP, Ahmed SF, Hughes IA. Consensus statement on management of intersex disorders. International consensus conference on intersex. *Pediatrics* 2006;118(2):e488-500.
2. Glassberg KI. The intersex infant: Early gender assignment and surgical reconstruction. *J Pediatr Adolesc Gynecol* 1998;11(3):151-4.
3. Grumbach MM, Conte FA, Hughes IA. Disorders of sex differentiation. In: Larsen PR, Kronenberg HM, Melmed S, Polonsky KS, editors. *Williams textbook of endocrinology* (10th ed.). Philadelphia: W.B. Saunders; 2002.
4. Money J, Ehrhardt A. *Man & woman boy & girl: The differentiation and dimorphism of gender identity from conception to maturity*. Baltimore: Johns Hopkins University Press; 1972.
5. Money J, Hampson JG, Hampson JL. Hermaphroditism: Recommendations concerning assignment of sex, change of sex, and psychologic management. *Bull Johns Hopkins Hosp* 1955;97(4):284-300.
6. Crouch NS, Creighton SM. Minimal surgical intervention in the management of intersex

- conditions. *J Pediatr Endocrinol Metab* 2004;17(12):1591-6.
7. Karkazis KA. Early genital surgery to remain controversial. *Pediatrics* 2006;118(2):814-5.
  8. Karkazis K. *Fixing sex: Intersex, medical authority, and lived experience*. Durham and London: Duke University Press; 2008.
  9. Sheridan SL, Harris RP, Woolf SH. Shared decision making about screening and chemoprevention. A suggested approach from the U.S. Preventive services task force. *Am J Prev Med* 2004;26(1):56-66.
  10. Woolf SH. The logic and limits of shared decision making. *J Urol* 2001;166(1):244-5.
  11. Charles C, Gafni A, Whelan T. Shared decision-making in the medical encounter: What does it mean? (or it takes at least two to tango). *Soc Sci Med* 1997;44(5):681-92.
  12. Charles C, Gafni A, Whelan T. Decision-making in the physician-patient encounter: Revisiting the shared treatment decision-making model. *Soc Sci Med* 1999;49(5):651-61.
  13. Coulter A. Shared decision-making: A summary and future issues. In: Maslin AM, Powles TJ, editors. *Breast cancer: Sharing the decision* Oxford: Oxford University Press; 1999.
  14. Emanuel EJ, Emanuel LL. Four models of the physician-patient relationship. *JAMA* 1992;267(16):2221-6.
  15. Carlet J, Thijs LG, Antonelli M, Cassell J, Cox P, Hill N, et al. Challenges in end-of-life care in the ICU. Statement of the 5th international consensus conference in critical care: Brussels, Belgium, April 2003. *Intensive Care Med* 2004;30(5):770-84.
  16. Urowitz S, Deber R. How consumerist do people want to be? Preferred role in decision-making of individuals with HIV/AIDS. *Health Policy* 2008;3(3):e168-82.
  17. O'Neal EL, Adams JR, McHugo GJ, Van Citters AD, Drake RE, Bartels SJ. Preferences of older and younger adults with serious mental illness for involvement in decision-making in medical and psychiatric settings. *Am J Geriatr Psychiatry* 2008;16(10):826-33.
  18. Pieterse AH, Baas-Thijssen MC, Marijnen CA, Stiggelbout AM. Clinician and cancer patient views on patient participation in treatment decision-making: A quantitative and qualitative exploration. *Br J Cancer* 2008;99(6):875-82.
  19. Heyland DK, Cook DJ, Rocker GM, Dodek PM, Kutsogiannis DJ, Peters S, et al. Decision-making in the ICU: Perspectives of the substitute decision-maker. *Intensive Care Med* 2003;29(1):75-82.
  20. Roupie E, Santin A, Boulme R, Wartel JS, Lepage E, Lemaire F, et al. Patients' preferences concerning medical information and surrogacy: Results of a prospective study in a French emergency department. *Intensive Care Med* 2000;26(1):52-6.
  21. Kon AA. Assessment of physician directiveness: Using hypoplastic left heart syndrome as a model. *J Perinatol* 2004;24(8):500-4.
  22. Kon AA, Ackerson L, Lo B. How pediatricians counsel parents when no "Best-choice" Management exists: Lessons to be learned from hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med* 2004;158(5):436-41.
  23. Frosch DL, Kaplan RM. Shared decision making in clinical medicine: Past research and future directions. *Am J Prev Med* 1999;17(4):285-94.
  24. Woolf SH. Shared decision-making: The case for letting patients decide which choice is best. *J Fam Pract* 1997;45(3):205-8.
  25. In; *Canterbury v. Spence*, 464 F.2d 772 (D.C. Cir. 1972).
  26. American Academy of Pediatrics Committee on Bioethics. Informed consent, parental permission, and assent in pediatric practice. *Pediatrics* 1995;95:314-317.
  27. Kon AA. Healthcare providers must offer palliative treatment to parents of neonates with hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med* 2008;162(9):844-8.
  28. Towle A, Godolphin W. Framework for teaching and learning informed shared decision making. *BMJ* 1999;319(7212):766-71.
  29. Purves JT, Miles-Thomas J, Migeon CJ, Gearhart JP. Complete androgen insensitivity: The role of the surgeon. *J Urol* 2008;180:1716-1719.
  30. Guerra-Junior G, Maciel-Guerra AT. The role of the pediatrician in the management of children with genital ambiguities. *J Pediatr (Rio J)* 2007;83(5 Suppl):S184-91.
  31. Haidet P, Fecile ML, West H, Teal C. Reconsidering the team concept: Educational implications for patient-centered care *Patient Educ Couns* In press.
  32. Epstein R, Street R. *Patient-centered communication in cancer care: Promoting healing and reducing suffering*. Bethesda, MD: National Cancer Institute; 2007.
  33. Fallowfield L, Lipkin M, Hall A. Teaching senior oncologists communication skills: Results from phase I of a comprehensive longitudinal program in the United Kingdom. *J Clin Oncol* 1998;16(5):1961-8.
  34. Hoffman M, Steinberg M. Development and implementation of a curriculum in communication skills and psycho-oncology for medical oncology fellows. *J Cancer Educ* 2002;17(4):196-200.
  35. Lorin S, Rho L, Wisnivesky JP, Nierman DM. Improving medical student intensive care unit communication skills: A novel educational initiative using standardized family members. *Crit Care Med* 2006;34(9):2386-91.
  36. Suiptot S, Bonnaud-Antignac A. Using simulated interviews to teach junior medical students to disclose the diagnosis of cancer. *J Cancer Educ* 2008;23:102-7.
  37. Street RL, Jr. Information-giving in medical

- consultations: The influence of patients' communicative styles and personal characteristics. *Soc Sci Med* 1991;32(5):541-8.
38. Strull WM, Lo B, Charles G. Do patients want to participate in medical decision making? *JAMA* 1984;252(21):2990-4.
  39. Bruera E, Willey JS, Palmer JL, Rosales M. Treatment decisions for breast carcinoma: Patient preferences and physician perceptions. *Cancer* 2002;94(7):2076-80.
  40. Degner LF, Kristjanson LJ, Bowman D, Sloan JA, Carriere KC, O'Neil J, et al. Information needs and decisional preferences in women with breast cancer. *JAMA* 1997;277(18):1485-92.
  41. Marsh JL. To cut or not to cut? A surgeon's perspective on surgically shaping children. In: Parens E, editor. *Surgically shaping children: Technology, ethics, and the pursuit of normality*. Baltimore, MD: The Johns Hopkins University Press; 2006. p. 113-124.
  42. Azoulay E, Pochard F, Chevret S, Adrie C, Annane D, Bleichner G, et al. Half the family members of intensive care unit patients do not want to share in the decision-making process: A study in 78 french intensive care units. *Crit Care Med* 2004;32(9):1832-8.
  43. Ong LM, Visser MR, Lammes FB, de Haes JC. Doctor-patient communication and cancer patients' quality of life and satisfaction. *Patient Educ Couns* 2000;41(2):145-56.
  44. Orsino A, Cameron JI, Seidl M, Mendelssohn D, Stewart DE. Medical decision-making and information needs in end-stage renal disease patients. *Gen Hosp Psychiatry* 2003;25(5):324-31.
  45. Salkeld G, Solomon M, Short L, Butow PN. A matter of trust--patient's views on decision-making in colorectal cancer. *Health Expect* 2004;7(2):104-14.
  46. Tamar-Mattis A. Exceptions to the rule: Curing the law's failure to protect intersex infants. *Berkeley Journal of Gender, Law & Justice* 2006;59.
  47. Tamar-Mattis A, Diamond M. Managing variation in sex development. *J Pediatr Endocrinol Metab* 2007;20(4):552-3.
  48. Kon AA. Silent decisions or veiled paternalism? Physicians are not experts in judging character. *Am J Bioeth* 2007;7(7):40-2.
  49. Dayner JE, Lee PA, Houk CP. Medical treatment of intersex: Parental perspectives. *J Urol* 2004;172(4 Pt 2):1762-5; discussion 1765.
  50. Feder EK. Doctor's orders: Parents and intersexed children. In: Feder E, Kittay EF, editors. *The subject of care: Feminist perspectives on dependency*. Lanham, MD: Rowman and Littlefield; 2002. p. 294-320.
  51. Brosig CL, Whitstone BN, Frommelt MA, Frisbee SJ, Leuthner SR. Psychological distress in parents of children with severe congenital heart disease: The impact of prenatal versus postnatal diagnosis. *J Perinatol* 2007;27(11):687-92.
  52. Rosenthal ET, Biesecker LG, Biesecker BB. Parental attitudes toward a diagnosis in children with unidentified multiple congenital anomaly syndromes. *Am J Med Genet* 2001;103(2):106-14.
  53. Ryan S. Telling parents their child has severe congenital anomalies. *Postgrad Med J* 1995;71(839):529-33.
  54. Howe EG. Intersexuality: What should careproviders do now. *J Clin Ethics* 1998;9(4):337-44.
  55. Schober JM. Early feminizing genitoplasty or watchful waiting. *J Pediatr Adolesc Gynecol* 1998;11(3):154-6.
  56. Grewal D, Davidson HA. Emotional intelligence and graduate medical education. *JAMA* 2008;300(10):1200-2.
  57. Ross LF. Why "Doctor, if this were your child, what would you do?" deserves an answer. *J Clin Ethics* 2003;14(1-2):59-62.
  58. Ruddick W. Questions parents should resist. In: Kopelman L, Moskop J, editors. *Children and health care: Moral and social issues*. Dordrecht, the Netherlands: Kluwer Academic; 1989. p. 221-229.
  59. Truog RD. "Doctor, if this were your child, what would you do?" *Pediatrics* 1999;103(1):153-4.
  60. Braddock CH, 3rd, Edwards KA, Hasenberg NM, Laidley TL, Levinson W. Informed decision making in outpatient practice: Time to get back to basics. *JAMA* 1999;282(24):2313-20.
  61. Entwistle VA, Sheldon TA, Sowden A, Watt IS. Evidence-informed patient choice. Practical issues of involving patients in decisions about health care technologies. *Int J Technol Assess Health Care* 1998;14(2):212-25.
  62. Holmes-Rovner M, Llewellyn-Thomas H, Entwistle V, Coulter A, O'Connor A, Rovner DR. Patient choice modules for summaries of clinical effectiveness: A proposal. *BMJ* 2001;322(7287):664-7.
  63. Walter LC, Covinsky KE. Cancer screening in elderly patients: A framework for individualized decision making. *JAMA* 2001;285(21):2750-6.
  64. Elder JS, Peters CA, Arant BS, Jr., Ewalt DH, Hawtrey CE, Hurwitz RS, et al. Pediatric vesicoureteral reflux guidelines panel summary report on the management of primary vesicoureteral reflux in children. *J Urol* 1997;157(5):1846-51.
  65. Elwyn G, Edwards A, Kinnersley P, Grol R. Shared decision making and the concept of equipoise: The competences of involving patients in healthcare choices. *Br J Gen Pract* 2000;50(460):892-9.

**TABLE 1**

What do Parents Need to Know? Information that Should be Part of SDM for Genitoplasty  
in Pediatric Patients with DSD

<p><u>Diagnosis</u></p> <ul style="list-style-type: none"> <li>• Full details of the child's diagnosis, including chromosomes, internal and external organs, and possible atypical hormone exposure before birth.</li> </ul>
<p><u>Prognosis</u></p> <ul style="list-style-type: none"> <li>• Potential for fertility, together with options for assisted reproduction that exist now or seem likely during the child's reproductive lifetime;</li> <li>• Potential for sexual function, including the capacity for sexual pleasure, orgasm, and emotional fulfillment as well as for penetration;</li> <li>• Gender identity outcomes in children with the same condition;<sup>1</sup></li> <li>• Psychological outcomes in children with the same or similar conditions, to the extent data are available.</li> </ul>
<p><u>Rationale for Proposed Procedure(s)</u></p> <ul style="list-style-type: none"> <li>• Information about the purpose of proposed interventions (e.g., to alter appearance, to allow voiding of urine, to allow standing urination, to allow for future menstrual flow, to allow for future sexual intercourse, to support gender identity development);</li> <li>• Where multiple procedures are proposed, the rationale for each must be disclosed;</li> <li>• Where outcome data in support of the rationale are lacking, it is important to clarify the basis for assertions made to parents (i.e., provider opinion or experience).</li> </ul>
<p><u>Risks and Benefits of Proposed Procedures</u></p> <ul style="list-style-type: none"> <li>• All material risks of proposed procedures, including minor risks if are common, and remote risks if potential harm is serious;</li> <li>• Potential benefits;</li> </ul>

- The likelihood of various outcomes, where this is known, or the uncertainty of suggested outcomes;
- Psychological risks (e.g., if a surgery or its possible complications will increase the necessity for multiple genital exams in childhood, the risk for psychological harm from such exams);<sup>1</sup>
- Risks and benefits throughout the patient's lifespan, including known or suspected risk of damaging adult sexual sensation and the risk of psychological harm if the patient later regrets the decision;<sup>2</sup>
- Parents' false assumptions should be corrected (i.e., that genitoplasty is "reversible," that early genitoplasty will ensure adoption of the assigned gender or development of a heterosexual identity,<sup>1</sup> or that surgery will obviate the need to tell the child of his or her condition).

#### Alternatives to Proposed Treatment

- Alternative procedures, along with their risks and benefits as described above;
- Non-treatment, watchful waiting and delayed treatment, along with their risks and benefits;
- Non-surgical treatment options, including psychological counseling for the child and family.
- Parents need to know that children born with DSD may enjoy healthy relationships and a well-adjusted childhood without surgical treatment.<sup>3</sup>
- Alternative treatments must be disclosed, regardless of the patient's ability to pay or the availability of insurance coverage for alternatives. Where insurance may cover a procedure in childhood but not in adulthood, this possibility should be discussed.
- Parents should be informed of their right to receive a second opinion and their right to change clinicians if disagreements cannot be resolved.

#### Post-surgical Care

- Expectations for aftercare, including length of hospital stay, need for pain control, and potential

for post-surgical complications in the near and long term.

- Where the need for ongoing catheterization or vaginal dilation is a possibility: associated challenges and potential for psychological harm from these procedures. If the parents will be expected to perform the dilations or catheterization, they should know this in advance.

#### Clinician Experience

- Surgeons' level of experience with the proposed procedures along with the surgeons' and hospital's outcomes including types and rates of complication.
- Where there is evidence of better outcomes at centers of excellence, this information must be disclosed.<sup>4</sup>

#### Referrals to Professional and Peer Support

- Where decision-making is based on speculation about the child's future quality of life, such access may provide information critical for informed decision making.

#### Special Cases

##### *Where Vaginoplasty is Contemplated*

- Lifelong need for dilation;
- Risk of scarring that may preclude intercourse and diminish fertility;<sup>6</sup>
- Risk of psychological harm from vaginal dilation in childhood;<sup>1</sup>
- Patient groups have reported that the available techniques for vaginoplasty (i.e., self-dilation, skin substitution, and bowel vaginoplasty) have varying advantages and drawbacks across the lifespan. Adults who have made choices for themselves about these procedures have demonstrated that idiosyncratic personal preferences play a big role decision making. Parents need to understand these variables when making decisions about vaginoplasty on behalf of a child.

##### *Where Clitoroplasty is Contemplated*

- That long-term effects of nerve-sparing techniques on clitoral sensitivity and sexual function are unknown;
- Potential negative impact of reduced clitoral sensitivity on future sexual activity and intimate relationships.<sup>6</sup>

#### *Hypospadias Repair*

- Possibility of need for multiple surgery and risk of “hypospadias cripple”;<sup>7</sup>
- Risk for complications that would make repeat surgeries non-elective, and of the associated psychological risks if multiple genital surgeries in childhood should become necessary;<sup>8,9</sup>
- Where hypospadias repair is contemplated for the purpose of improving fertility: other options for addressing impaired fertility (i.e., alternative insemination);
- Where hypospadias repair is recommended to increase the probability that the child will be able to stand for urination: possibility that the child will be able to stand to urinate without the repair, and the possibility of fanning or spraying of the urine stream after repair which could make standing urination difficult.<sup>8</sup>

#### *The Older Pediatric Patient*

- As the child with a DSD matures, it is appropriate to involve him or her in decisions about ongoing treatment. The AAP provides guidance regarding assent for procedures that are elective.<sup>5</sup> There is uncertainty about the validity of parental consent alone for genitoplasty or gonadectomy at any age; it is clearly inappropriate, and possibly a violation of civil rights, to perform these procedures on an older child or teen without assent;
- When the patient reaches the age of 18, he or she will have the right to access the entire medical record. Parents who are considering limiting disclosure to the child need to be aware of this;
- Where state or constitutional law accords mature minors the authority to make their own informed choices, the information necessary to make an informed decision must be fully provided to the minor in understandable language;

- Where a mature minor and a parent disagree over treatment choices, or where disagreement over treatment arises between parents after the SDM process, the provider should obtain ethical and legal advice before treatment is performed.

### Specific Diagnoses

#### *Androgen Insensitivity Syndrome*

- Use of misleading terminology (i.e., referring to testes as “twisted ovaries” or referring to an XY genotype as “female chromosomes”) is contrary to principles of informed consent and is ethically inappropriate;
- If reproductive structures such as a uterus or testes are unexpectedly discovered during surgery, separate informed consent and possibly a court order should be obtained before any resection.

#### *Congenital Adrenal Hyperplasia*

- Percentage of 46,XX children with CAH who ultimately develop a male gender identity;<sup>1</sup>
- Where a “one-stage repair” is contemplated: the distinct procedures involved (i.e., vaginoplasty and clitoroplasty), and the differing rationales for each;
- Where vaginoplasty is recommended for functional reasons: whether the functional issue is immediate (i.e., to allow for urine flow) or anticipated in the future (i.e., to avoid possible UTIs; to allow for eventual menstrual flow; to allow for future heterosexual intercourse).
- Where genital surgery is recommended to prevent possible UTIs due to presence of a urogenital sinus: the absence of evidence that UTIs are more common in girls with a urogenital sinus, and the possibility that surgery could increase the incidence of UTIs.<sup>10</sup>

### Pitfalls to Avoid

#### *Failure to Clarify Quality of Evidence*

- Offering information without clarifying which assertions are based on provider’s opinion, rather than evidence-based published studies;

- Failing to explain existence of medical controversy about treatment options or existence of conflicting data.

#### *Unwarranted Pressure*

- Urging parents to decide quickly when the condition is in fact non-emergent;
- Offering biased information (i.e., urging a reluctant parent to consent to genitoplasty in order to prevent urinary tract infections when in fact the risk of UTI is minor, but the provider is convinced that genitoplasty is necessary for psychological functioning or parental comfort);
- Threats to involve child protective services (It is highly unlikely that state authorities will intervene just because a parent declines to authorize elective surgery.);
- Suggestions that a child will commit suicide if surgery is declined, in the absence of evidence for such a claim.

#### *Excessive Optimism*

- Excessive optimism about functional and cosmetic outcomes of genitoplasty;
- Putting a “spin” on factual data. For example:
  - A statement such as “In favorable cases, the maximum number of operations can be two or three,” could leave a parent with the impression that the maximum number of operations *will* be two or three;
  - A statement such as “it is possible to achieve both favorable functional and favorable cosmetic outcomes,” without qualification, may leave parents with the impression that such an outcome is likely or even assured.<sup>10</sup>

#### *Practicing Outside the Field of Competency*

- If a surgeon with no specialized training in child psychology, child development or a related field recommends genitoplasty for reasons of ensuring normal sexual development, this could be found to be practicing outside the field of competency. In such a case, the physician would be held to the standard of care of a specialist in the field.<sup>11</sup> Consultation with an appropriate

specialist is recommended if surgery is contemplated for psychosocial reasons.

## REFERENCES

1. Lee PA, Houk CP, Ahmed SF, Hughes IA. Consensus statement on management of intersex disorders. International consensus conference on intersex. *Pediatrics* 2006;118(2):e488-500.
2. Tamar-Mattis A. Exceptions to the rule: Curing the law's failure to protect intersex infants. *Berkeley Journal of Gender, Law & Justice* 2006;59.
3. Consortium on Disorders of Sex Development. Clinical guidelines. 2005.
4. Johnson v. Kokemoor, 545 N.W.2d 495 (Wis. 1996).
5. American Academy of Pediatrics Committee on Bioethics. Informed consent, parental permission, and assent in pediatric practice. *Pediatrics* 1995;95:314-317.
6. Arlt W, Krone N. Adult consequences of congenital adrenal hyperplasia. *Horm Res* 2007;68 Suppl 5:158-64.
7. Stecker JF, Jr., Horton CE, Devine CJ, Jr., McCraw JB. Hypospadias cripples. *Urol Clin North Am* 1981;8(3):539-44.
8. Bubanj TB, Perovic SV, Milicevic RM, Jovicic SB, Marjanovic ZO, Djordjevic MM. Sexual behavior and sexual function of adults after hypospadias surgery: A comparative study. *J Urol* 2004;171(5):1876-9.
9. Migeon CJ, Wisniewski AB, Gearhart JP, Meyer-Bahlburg HF, Rock JA, Brown TR, et al. Ambiguous genitalia with perineoscrotal hypospadias in 46,XY individuals: Long-term medical, surgical, and psychosexual outcome. *Pediatrics* 2002;110(3):e31.
10. Nabhan ZM, Rink RC, Eugster EA. Urinary tract infections in children with congenital adrenal hyperplasia. *J Pediatr Endocrinol Metab* 2006;19(6):815-20.
11. Rivkees SA. It is (sort of) a boy and (sort of) a girl. You have (sort of) a say and you (sort of) don't? The uneasiness of genital restoration surgery. *J Pediatr Endocrinol Metab* 2006;19(11):1285-9.
12. Carrasco v. Bankoff, 220 Cal. App. 2d 230 (1963).